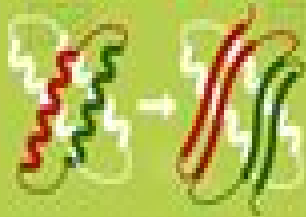


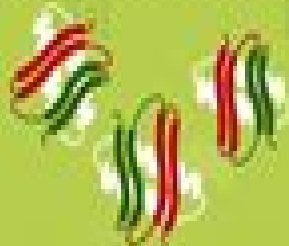
PATHOPHYSIOLOGY OF PRION DISEASE AND ITS EFFECT ON NEURAL SYSTEM



Prion disease belong to the family of disease designated as protein misfolding-related diseases.



In prion diseases, the alpha-helical PrP^C is converted into its misfolded and beta sheet form, PrP^{Sc}.



The absence of normal protein renders the cell non-functional. It also leads to more PrP^C conversion to PrP^{Sc}.



The accumulation of PrP^{Sc} hampers the neuron's Unfolded Protein Response (UPR) that regulates the production of normal proteins and causes neural loss and degeneration.



The superficial features of prion disease resemble other neurodegenerative disorders: Alzheimer's disease (AD), Parkinson's disease (PD) and Amyotrophic Lateral Sclerosis (ALS)

Prion Biology And Diseases

IM Harris



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